Paroxysmal dystonic choreoathetosis

D. Rogers

Burden Neurological Hospital, Bristol, UK

A 30-year-old man with mild learning disability (IQ:72) and cerebral palsy with mild generalized choreoathetosis was referred with paroxysmal motor disorder and episodic depressive mood disorder. Aged 11 months, he had developed episodes of alternating flaccid hemiparesis followed by episodes when he was unable to move or talk but remaining conscious lasting from several minutes to longer. Episodes could be precipitated by excitement such as opening Christmas presents or, later, his favourite football teams scoring a goal or losing. His longest interictal period had been 4 weeks. There was a family history of epilepsy. His EEG aged 6 showed paroxysmal spike and slow waves but no change during episodes of weakness. There was no improvement with phenobarbitone or phenytoin. Aged 11, oculogyric crises started during episodes. Aged 19, the EEG showed photosensitivity but was otherwise normal. Episodes were made worse with Sinemet or amphetamines. From age 20, he regularly needed 2 to 5 days a month off work because of episodes. From age 27, episodes could be followed by depression lasting days to months, including suicidal attempts such as swallowing bleach aged 29. Once started on carbamazepine 400 mgs daily, he still had episodes every 2 weeks but has not needed any further time off work, has had no further depressive episodes and is now able to live independently. He could still have rare episodes lasting up to 3 days such as when England won at Wembley.

Photosensitivity in juvenile myoclonic epilepsy

Margaret Beirne, Barbara Acomb & Richard Appleton

The Roald Dahl EEG Unit, Alder Hey Children's Hospital, Liverpool, UK

Photosensitivity (PS) is reported to occur in 60–70% of patients with juvenile myoclonic epilepsy (JME). The incidence may be higher, possibly reflecting the duration of intermittent photic stimulation (IPS), the technique used to provoke PS in the EEG laboratory.

A 2-year retrospective review of EEGs was undertaken on all children with JME. JME was diagnosed on clinical (age of onset 6–16 years; myoclonic seizures; at least one tonic–clonic seizure; no additional neurological problems) and EEG (generalized spike and wave or polyspikes) criteria. IPS was based on international guidelines using flash rates of 1–60 per second but undertaken over a period of 5 minutes. Fifty-six patients (31 female) with a median age of 13 years (range: 7–16) were identified. No patient was receiving an antiepileptic drug at the time of their initial EEG. Fifty-five patients (99%) were photosensitive, of whom 37 (67%) demonstrated a photoparoxysmal response and another 16 (29%) had a photoconvulsive (myoclonic) response; no patient experienced a tonic–clonic seizure. Eighteen of the 55 patients (33%) manifest PS only after 4 minutes of IPS.

These results suggest that the incidence of PS in JME may be higher than previously reported. Although IPS was more prolonged in this study than in most laboratories and the implications of a photically-induced seizure are greater for an adult than a child/teenager, the identification of PS is important because of counselling and treatment implications.

An audit of the appropriateness of EEG requests in a District Hospital setting

D. Smith, R. Bartolo & B. Tedman

The Walton Centre for Neurology and Neurosurgery, Lower Lane, Fazakerley, L9 7LJ, UK

This project included retrospective and prospective components. In each the case notes and EEG reports of patients undergoing EEGs at the Wrexham Maelor Hospital were examined with data collected on the source of and reason for request, result, clinical diagnosis and role of EEG in establishing diagnosis. For comparison, the same process was conducted at the Regional Neurology Centre.

Retrospective analysis revealed that the EEG was being used as a diagnostic tool particularly for the 'exclusion of epilepsy' in patients with 'funny turns' where it is more likely to yield potentially misleading than diagnostic information. The role of the EEG was categorized as 'influencing management' (14.2%), 'justifiable' (23%) and 'inappropriate' (62.8%). The proportion of inappropriate requests in a General Neurology Clinic and a Regional Epilepsy Clinic were 25.6% and 11.2%, respectively.

An 'Inappropriateness' standard of 35–40% was agreed and guidelines were issued to facilitate achievement thereof. In the prospective study there was a 25% reduction in the total number of EEGs and a 50% reduction in 'diagnostic EEG' in patients with 'funny turns'. The proportion of inappropriate requests fell to 39% while those influencing management rose to 27%. A standardized request form, based on the guidelines, is now in use and the reduction in EEG requests has been maintained for 2 years.

This successful audit has potential benefits for both patients and clinicians within the setting of a more comprehensive local Neurophysiology Service.